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Case report

Thoracic wall metastases from nasopharyngeal carcinoma in a teenage boy: A case report

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ABSTRACT

We report an extremely rare case of nasopharyngeal carcinoma in a 14-year old boy presenting with neck and chest wall metastasis. Radiological and histological examinations established the diagnosis of nasopharyngeal carcinoma with non-keratinizing undifferentiated, WHO Type III form. He received 3 courses of chemotherapy followed by 35 cycles of radiotherapy. The primary site and the neck and chest wall metastases responded well with treatment. At 3 years follow up, the patient is still in a symptom-free and relapse-free state. Despite presenting late, juvenile onset WHO type 3 NPC showed a good response to chemoradiotherapy in our teenage patient.

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1. Introduction

In worldwide population, the incidence of nasopharyngeal carcinoma (NPC) had 2 peaks: the initial peak is in the late adolescence/early adulthood and a later peak is later in life (above 65 years). NPC is uncommon in children and teenage group. It accounts for around 1–5% of all primary cancers in children. ²

The commonest place for distant metastases from NPC is to the bone, followed by the lung and liver.³ Bony metastatic sites are usually scattered and multifocal. A search of literature revealed only a few cases that have reported sternal and rib metastasis in adults,^{3–5} but none of them occurred in young patients. We report a unique case of a teenager with thoracic wall metastases from NPC.

2. Case report

A 14-year old teenager presented with bilateral enlarging neck swelling of 5 months duration which had not responded to multiple different courses of oral antibiotics prescribed by his doctors. The swelling became progressively enlarged with time but relatively painless. It did not interfere with his breathing and oral

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intake. There were no nasal and ear symptoms complaint, but he reported loss of appetite and weight significantly for the past 3 months. He had intermittent vague, ill-defined left sided chest pain for 1 month which had been treated by different doctors as gastritis, costochondritis and was even investigated for acute coronary syndrome but was never concluded.

On examination of the neck, there were hard, fixed bilateral cervical lymph nodes at levels II–IV measuring $8\ cm \times 6\ cm$ in the greatest dimension. Rigid nasoendoscopy revealed bilateral nasopharyngeal mass with irregular surface. The mass obliterated both Fossa of Rosenmuler and obscured the Eustacian tubes, with the left mass slightly bigger and more prominent than the right. Chest examination reveals no abnormal findings apart from vague tenderness on palpation and percussion all over the left thoracic wall.

An ultrasound scan of the neck revealed a large complex mass featuring cystic and solid component with minimal colour doppler signal seen within. There was no vascular compromise in the neck. Fine needle aspiration cytology (FNAC) of the swelling reported atypical cells and was negative for tuberculosis.

CT scan of the base of skull until thorax showed ill-defined mass involving bilateral Fossa of Rossenmuller and retropharyngeal space measuring $3.0~{\rm cm}\times 6.0~{\rm cm}$ with necrotic changes (Fig. 1). Multiple enlarged cervical, submandibular and supraclavicular lymph nodes were seen bilaterally with some central necrosis. The largest node was $5.9~{\rm cm}\times 6.1~{\rm cm}\times 7.7~{\rm cm}$, at the left neck posterior triangle (Fig. 2). There was no clear demarcation between those lesions and the adjacent muscles. Both internal jugular veins

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Fig. 1. Ill-defined mass involving bilateral Fossa of Rossenmuller and retropharyngeal space with necrotic changes (arrows).

were compressed by the enlarged lymph nodes. CT thorax highlighted a lobulated lesion over the left lateral chest wall measuring 5.6 cm \times 8.3 cm \times 5.6 cm with a necrotic center (Fig. 3). There was no clear plane between the lesion and its adjacent pleural and thoracic wall soft tissue. Destruction of the left lateral 5th rib was evident.

He underwent endoscopic biopsy of the nasopharyngeal mass and a repeat needle aspiration cytology of the neck under general anaesthesia. A CT guided transthoracic needle biopsy of the left chest wall lesion was also done concurrently. Histopathologic examination reported as non keratinizing undifferentiated, WHO Type 3 nasopharyngeal carcinoma and malignant infiltration of the neck and thoracic wall by the similar tumor.

The patient was subsequently referred to a tertiary center with oncology facility where he received 3 cycles of chemotherapy (cisplatin and 5-flourouracil) and 30 cycles of radiotherapy. The primary site and neck metastases responded well with treatment. The chest wall tumor shrunk considerably and at the last follow up after three years of diagnosis, the patient was still in a



Fig. 3. CT thorax showed a lobulated lesion over the left lateral chest wall measuring $5.6~\text{cm} \times 8.3~\text{cm} \times 5.6~\text{cm}$ with a necrotic center (arrow) and destruction of the left lateral 5th rib.

symptom-free and relapse-free state. He is still under a three-monthly follow-up in our outpatient clinic.

3. Discussion

A search in the literature revealed that most of the chest wall metastasis was originated from the tumor in adjacent structures: the lungs, pleura, mediastinum and breasts. Primary lung malignancies that involve chest wall spread via mediastinal, intrapulmonary and hilar lymphatic drainage. Lung metastasis is a known complication of advanced nasopharyngeal carcinoma. However, in our case, no lung parenchyma or intrathoracic lesions were detected. Thus, this could still be a separate pathology from the nasopharyngeal lesion. Hence, we needed to establish the diagnosis and the transthoracic biopsy confirmed the diagnosis of chest wall metastasis from nasopharyngeal carcinoma.

It is imperative to recognize the signs and symptoms of nasopharyngeal carcinoma and the fact that NPC can occur in pediatric patients, especially in a highly prevalent South Asian population. Suspicious cases attended by the primary care doctors should promptly be referred to ENT specialty clinic for screening and further work-up. Our patient had ill-defined left sided chest pain which had garnered various diagnoses but never resolved. Bone destruction by tumor infiltration as seen in our patient are known to cause dull, vague and ill defined pain until a pathological fracture occurs; the pain then becomes sharp and localized. The long lasting dull bony pain is conducted by unmyelinated fibers in bone

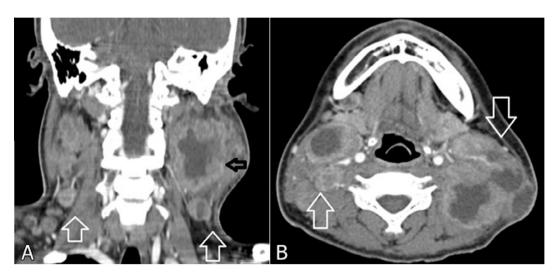


Fig. 2. (A) Axial CT picture showing multiple enlarged cervical, submandibular and supraclavicular lymph nodes bilaterally with some central necrosis (arrows). (B) Coronal CT picture showing bilateral multiple nodes (white arrows) with the largest node measuring 5.9 cm × 6.1 cm × 7.7 cm, at the left neck posterior triangle (dark arrow).

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marrows and the quality of the pain becomes more localized and sharp as the malignancy involves the highly pain-sensitive, nociceptors-rich periosteum layer.

Lim et al. reported that majority of the pediatric and adolescent nasopharyngeal carcinoma presented mainly with neck lumps, followed by headache and epistaxis. None of the patients in their series reportedly had a family history of nasopharyngeal carcinoma. Most of young patients with NPC presented at late stages of the disease. Chemoradiotherapy increased survival rates for these patient but they have high recurrence and distant metastasis rate of approximately 39%. ^{2,7-9} WHO Type III undifferentiated nasopharyngeal carcinoma is the most common type and have the best prognosis compared to other types as it responds better to radiotherapy and chemotherapy.

A study revealed that 5-year overall survival rate was 71% for juvenile nasopharyngeal carcinoma (jNPC) and 58% for adult nasopharyngeal carcinoma (aNPC).⁸ Relapse-free survival rate was also higher in jNPC group compared to the aNPC group. The jNPC patients had better survival than aNPC patients despite presenting at later stages.⁸ Early detection and treatment improves prognosis.

4. Conclusion

A rare case of thoracic wall metastases from NPC in a teenage boy is reported. Despite late and aggressive presentation, juvenile onset WHO type 3 NPC showed a good response to chemoradiotherapy in our case.

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